SHORT COMMUNICATION

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A Clinical Sign in Pediatric Sclerosis, Farhad's Sign

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ABSTRACT

The scleroderma group of diseases is characterized by the presence of hard skin. The pathogenesis of scleroderma include excessive production of collagen and extracellular matrix by fibroblasts, not only in the skin but also in vital organs and around blood vessels with endothelial cell injury. Systemic Sclerosis (SSc) usually constitutes less than 1% of most pediatric rheumatology clinic populations. Females are affected much more commonly than males.

Key words: pediatric systemic sclerosis, clinical sign.

The scleroderma group of diseases is characterized by the presence of hard skin. The pathogenesis of scleroderma include excessive production of collagen and extracellular matrix by fibroblasts, not only in the skin but also in vital organs and around blood vessels with endothelial cell injury (1). Systemic Sclerosis (SSc) usually constitutes less than 1% of most pediatric rheumatology clinic populations (2). Females are affected much more commonly than males.

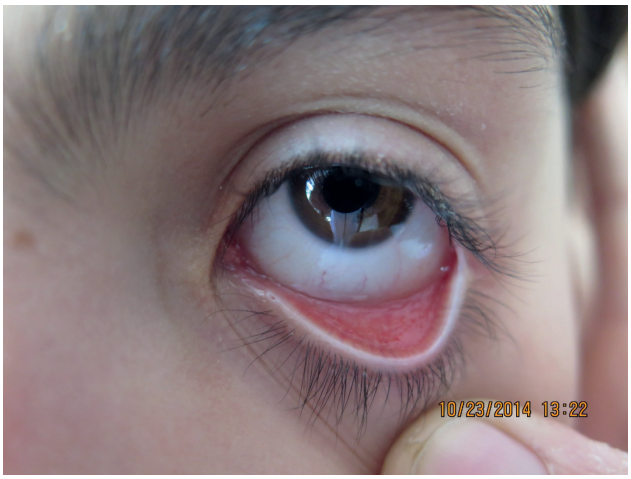
The earliest skin manifestation of SSc is nonpitting inflammatory edema of the hands, which results in restrictive range of motion. With time, this progress to skin thickening with an inability to lift the skin (3).

Progressive thickening and tightness of the skin result in joint contractures and an inability to fully open the mouth. Other skin manifestations include abnormal nail folds and calcinosis, telangiectasia, areas of hyperpigmentation and hypopigmentation (4).

Musculoskeletal involvement is common. Fibrosis of the joint capsule and thickening of the synovium results in joint contractures, a problem exacerbated by the thickened and tight skin around the joint.

Gastrointestinal involvement is a major cause of morbidity. The ma-

Figure 1. Patient with sclerodermica



Figrue 2. Patient with sclerodermia

jor current cause of mortality results from pulmonary involvement.

Renal disease, formerly the major cause of mortality, is now more manageable. The heart is also commonly affected.

Clinical methods to quantify skin thickening have been validated in adults but not in children (3).

Here is described very simple and useful sign to evaluate thickening and tightness of the skin in children as Farhad's sign:

In order to examine conjunctiva for anemia in children, we have to pull the lower eyelid downwardly. It is easily done in healthy people (5, 6) however; it wouldn't be done in patients with sclerodermia (Figure 1, 2)

This is a valuable finding that can be seen in all patients with systemic sclerosis. We name that Farhad's sign and use it in clinical examination of all suspected sclerodermia patients; and recommend it to all young physicians at practice.

CONFLICT OF INTEREST: NONE DECLARED.

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